

## SYPHILITIC AMYOTROPHY\*

### A CASE OCCURRING AFTER APPARENTLY SUCCESSFUL TREATMENT IN PRIMARY SYPHILIS

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Syphilitic amyotrophy, or muscular atrophy of syphilitic aetiology, was first described by Ballet (1894). The first case published in English was that of Mott (1910), who gave the history and clinical findings in a 36-year-old patient seen at the Charing Cross Hospital.

Muscular atrophy in syphilis is due to a progressive degeneration of the anterior horn cells in the spinal cord and may result from any variety of syphilis of the spinal cord, but the term syphilitic amyotrophy is usually reserved for the muscular atrophy which occurs in meningomyelitis. To clarify this statement the various types of syphilis of the spinal cord may be classified as follows:

- (1) *Parenchymatous*, i.e. *Tabes dorsalis*.
- (2) *Meningeal* (i) *Pachymeningitis*.  
(ii) *Vascular*.  
(iii) *Meningomyelitis*.

***Tabes Dorsalis*** Muscle wasting may occur in a small number of tabetics. It is usually confined to the lower limbs. Two cases of this nature were studied histologically by Wilson (1911), who observed degenerative changes in the anterior horn cells which he attributed to the action of syphilitic toxin as there was insufficient evidence of either peripheral neuritis or meningitis to account for them.

#### *Meningeal*

(i) *Pachymeningitis* is a gummatous condition, which may be:

- (a) Localized as a gumma as in meningitis serosa circumscripta,  
or
- (b) Diffuse as in pachymeningitis cervicalis hypertrophica.

Pachymeningitis causes nerve root and spinal cord compression. It may also cause occlusion of the spinal arteries resulting in infarction of the spinal

cord. These lead to degeneration of the anterior and posterior nerve roots of the affected segments. The patients present with root pains and paraesthesiae at the level of the lesion, followed by sensory loss and muscle atrophy. In pachymeningitis cervicalis hypertrophica there are in addition bulbar nerve lesions and there may be a Horner's syndrome. Lumbar puncture shows the presence of Froin's syndrome, that is, a manometric block and a high cerebrospinal fluid protein content.

(ii) *Vascular* A syphilitic arteritis may lead to an intravascular thrombosis and consequent infarction of the area of the cord supplied by that particular artery. This usually manifests itself dramatically with sudden onset of a flaccid paralysis of the legs with incontinence of urine and faeces. However, a lower motor neurone lesion of a single muscle or group of muscles, occurring as a result of a small infarct in the anterior horn, may be an isolated feature.

(iii) *Meningomyelitis* This occurs as a result of a chronic meningeal inflammation which damages the more peripheral fibres of the spinal cord. Its onset is insidious and its features include sensory loss, muscular atrophy, sphincter disturbances, and sometimes spasticity of the lower limbs. Amyotrophic meningomyelitis was very fully described by Martin (1925) who analysed the symptoms and signs in sixty cases, including seven of his own. In amyotrophic meningomyelitis there is bilateral or unilateral atonic atrophy beginning in the small muscles of the hands, in the shoulder girdle, or in the legs, and often preceded by pain. In contrast to the muscular atrophy of primary motor neurone disease, which is hypertonic due to associated anterolateral tract involvement, syphilitic muscular atrophy is atonic and the deep reflexes are abolished. All of the patients reviewed by Martin showed muscle wasting, but only 77 per cent. presented on

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account of muscle weakness; the rest presented with pain which was usually constant in character. In 40 per cent. of the series the pupils were normal, 28 per cent. showed true Argyll Robertson pupils, and the remainder either a sluggish reaction to light or pupillary irregularities.

Two of Martin's cases came to *post mortem* and were examined histologically. The findings were:

- (1) Leptomeningitis.
- (2) Marginal degeneration of the white matter of the spinal cord.
- (3) Very severe destruction of the cells of the grey matter of the cervical enlargement.
- (4) Arteritis of the meningeal and intraspinal arteries.

Four patients that Martin treated showed a halting of the disease process and one further patient showed a regression of the amyotrophy after treatment.

The following case is presented as it shows the occurrence of syphilitic amyotrophy of Martin's type within 10 months of apparently adequate antisypilitic treatment with satisfactory serological improvement.

#### Case History

**A 37-year-old Canadian accountant** gave a history of acute urethral gonorrhoea in 1943 and 1959 (the serology at this time was known to be negative) but of no other illnesses. He presented at the West London Hospital in October, 1962, with a history of having been treated for a "rectal abscess" by his general practitioner some 3 months previously with Graneodin (Neomycin and Gramicidin) ointment but no other medication. He denied any sexual exposure over the preceding year but gave a history of intimate contact with a male acquaintance a week or so before the appearance of the abscess. He stated that at this time he was a regular blood donor but subsequent inquiries to the National Blood Transfusion Service have failed to confirm this statement.

**Examination** He was a healthy-looking man of average build and the only abnormal findings were bilaterally enlarged inguinal glands and a small ulcer about 1 in. (2.5 cm.) posterior to the anus; dark-ground examination of serum from this was negative. The blood Wassermann reaction was strongly positive and the Kahn test positive.

**Diagnosis and Treatment** A diagnosis of sero-positive primary syphilis was made and treatment undertaken with procaine penicillin 900,000 units daily for 10 days. There was no Jarisch-Herxheimer reaction. 6 weeks after the end of the course of treatment the titre of the reagin tests was unchanged, but by 10 weeks the result of the Wassermann reaction was positive, rather than strongly positive, the titre of the Kahn test being unchanged.

He was then given a second course of procaine penicillin 900,000 units daily for 10 days and 3 weeks later the Wassermann reaction was reported as "anticomplementary" and the Kahn test as weakly positive.

**Progress** He was next seen in August, 1963, that is some 10 months after the initial course of antisypilitic treatment, when he complained of fleeting pains in both hands for the preceding 1 to 2 months and shooting cramp-like pains in the legs for 2 days. He steadfastly denied any sexual exposure.

**Examination** There was no sensory loss or change and there was no abnormality of the cranial nerves or of the tendon reflexes, the only abnormal finding being wasting of both hypotenar eminences with associated loss of power; otherwise examination gave normal results; the blood pressure was 104/85 mm. Hg. An x ray of the thoracic inlet and thoracic spine showed no significant abnormality. Lumbar puncture produced a clear fluid of which the protein content was 70 mg./100 ml. with a slight increase in globulin, and 2 mononuclear cells per c.mm., the Lange test and Wassermann reaction of the cerebrospinal fluid being negative. At this time the blood Wassermann reaction was negative and the Kahn test weakly positive.

**Diagnosis** Syphilitic amyotrophy was diagnosed and the patient was given a further course of procaine penicillin 900,000 units daily for 10 days.

**Progress** In January, 1964, he stated that he had been symptom-free since the conclusion of the course of treatment; there were no abnormal findings on physical examination, the muscle wasting having completely disappeared. At this time the Wassermann reaction was "anticomplementary" and the Kahn and Reiter protein complement-fixation tests weakly positive. This serological picture was repeated at four subsequent attendances during 1964 and 1965. Lumbar puncture in September, 1965, showed a clear acellular fluid with a protein content of 35 mg./100 ml. and no increase in globulin, the Lange test and Wassermann reaction being negative.

When he was last seen in May, 1966, the blood Wassermann reaction was "anticomplementary" and the Kahn and Reiter protein complement-fixation tests both weakly positive. The patient was sign- and symptom-free.

#### Discussion

Although syphilitic invasion of the nervous system is not uncommon, evidence of syphilitic invasion of the cerebrospinal fluid occurring in up to 25 per cent. of patients with syphilis (King and Nicol, 1964), overt disease which depends upon the immune status of the host is rare in early syphilis; indeed meningovascular and vascular syphilis was observed in only 3.3 per cent. of the male untreated cases of Boeck reviewed by Gjestland (1955). Hahn, Cutler, Curtis, Gammon, Heyman, Johnwick,

Stokes, Solomon, Thomas, Timberlake, Webster, and Gleeson (1956) reviewed 765 cases of asymptomatic neurosyphilis and determined the maximum cumulative probability of progression to symptomatic neurosyphilis, which they found to be:

0.55 per cent. after 1 yr  
1.69 per cent. after 3 yrs  
2.02 per cent. after 5 yrs  
3.31 per cent. after 7 yrs

Penicillin is the drug of choice in the treatment of syphilis and a continuous prolonged penicillinaemia rather than a high concentration is necessary for cure. King (1959) suggested the use of procaine penicillin 600,000 units daily for 10 days. Hellerström and Skog (1962) and Smith and Price (1957) demonstrated the efficacy of procaine penicillin; in the majority of their cases Hellerström and Skog used seventeen injections of 600,000 units procaine penicillin and saw no case of clinical relapse, the cerebrospinal fluid tests all being negative one year or more after treatment in the 51 patients in whom they were carried out. In the "Blue Star" investigation of Smith and Price (1957), only four patients out of 956 developed asymptomatic neurosyphilis.

Progression of neurosyphilis after treatment is by no means unknown, twelve of the 765 cases of asymptomatic neurosyphilis treated by Hahn and others (1956) showed possible progression, but they observed only one definite failure and this patient responded satisfactorily to re-treatment at a higher dosage. An instance of progression of symptomatic neurosyphilis, in this case general paresis, after 31,000,000 units procaine penicillin was described by Birkett (1961). Alergant (1965) reported the case of a man seen at the Liverpool Royal Infirmary with a relapse of secondary neurosyphilis, administration elsewhere 9 months earlier of penicillin in a sub-curative dose having masked the features of the condition until relapse occurred.

The present case of syphilitic amyotrophy of Martin's type occurring within a year of the apparently adequate treatment of early syphilis is unusual, and also illustrates Neisser's dictum that no one should be passed as cured of syphilis until the cerebrospinal fluid has been examined and found to be normal. Recent work by Lawton Smith and Israel (1967) using the fluorescent treponemal antibody test on cerebrospinal fluid suggests that the tests routinely employed may be inefficient in

detecting syphilitic invasion of the cerebrospinal fluid and that this may be more common than is generally recognized.

### Summary

A case is described of syphilitic amyotrophy in a 37-year-old man within a year of apparently successful treatment for primary syphilis. Most physicians would accept that a dosage of 18,000,000 units procaine penicillin, albeit in two courses, should cure early syphilis, but this case illustrates the necessity of prolonged clinical and serological follow-up and cerebrospinal fluid examination.

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### L'amyotrophie syphilitique

#### RÉSUMÉ

Un cas d'amyotrophie syphilitique survenu chez un homme de 37 ans moins d'un an après un traitement apparemment efficace pour la syphilis primaire est décrit. La plupart des spécialistes accepteraient qu'un dosage de 18,000,000 unités de procaine pénicilline, bien que donnés en deux séries, devrait guérir la syphilis primaire, mais ce cas démontre la nécessité de suivre de près et pendant longtemps le développement clinique et sérologique et de faire aussi l'analyse régulière du liquide céphalo-rachidien.